

# Proximally Wasting Away: A Case of Monoclonal Gammopathy of Renal Significance

Jaimie Barrera Gonzalez DO, Christine Palazzolo DO, Toni Sabbouh MD  
Pennsylvania Hospital of the University of Pennsylvania, Philadelphia, PA

## INTRODUCTION

- A monoclonal gammopathy of renal significance (MGRS) is a proliferative plasma-cell or B-cell disorder which results in pathologic kidney damage, but otherwise does not meet criteria for hematologic malignancy
- Common symptoms of MGRS include impaired kidney function, proteinuria, and microscopic hematuria
- Once renal biopsy confirms MGRS, further characterization is based upon on the region of injury along the nephron which can be glomerular, tubular, or vascular
- Below we report a case of MGRS light chain proximal tubulopathy (LCPT) with Fanconi Syndrome

## CASE PRESENTATION

- A 76-year-old female with hypertension, prediabetes, and glaucoma (on methazolamide) presented with nausea, vomiting, and poor intake
- Initial laboratory workup was significant for an acute kidney injury, non-anion gap metabolic acidosis (NAGMA), hypophosphatemia, and hypokalemia with urine studies suggesting renal wasting
- Initial management included discontinuing her home methazolamide and treating her nausea/vomiting
- Despite aggressive repletion, NAGMA persisted, and urine studies suggested a type 2 (proximal) renal tubular acidosis (RTA) with Fanconi syndrome
- Consequent paraprotein workup was significant for IgA kappa light chains
- She underwent a bone marrow biopsy which revealed 6% clonal plasma cells
- Confirmatory kidney biopsy was not pursued given patient preference, however MGRS LCPT was suspected

## DATA

142	112	21	110
2.9	17	1.44	

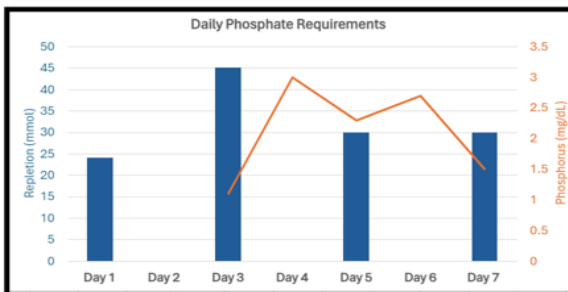
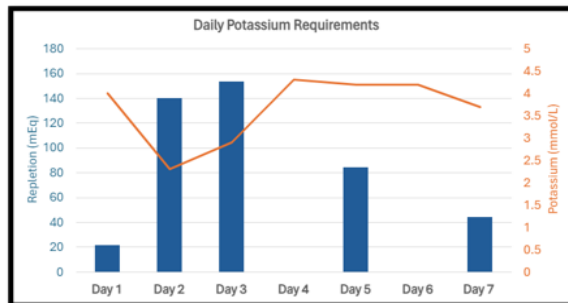
Mg 2.1  
P 1.1

**Figure 1.** Labs on day 3, despite electrolyte repletion

SPEP	IgA kappa band in $\beta$ region	
UPEP	Free kappa light chain	
Paraprotein workup		3.09

Ur K	90.5 mmol/L
Ur K:Ur Cr	19
Fe PO <sub>4</sub>	36%
Fe UA	15%
Ur Protein/Cr Ratio	3.57 g/g
Ur Albumin/Cr Ratio	0.3 g/g

**Table 2.** Proteinuria and renal wasting of potassium, phosphate, and uric acid demonstrated by urine potassium creatinine ratio >2.5% (Ur K:Ur Cr), fractional excretion of phosphate (Fe PO<sub>4</sub>) >3%, and fractional excretion of uric acid (Fe UA) >5%.



**Figure 2.** Potassium (top) and phosphate (bottom) levels with required daily repletion.

## DISCUSSION

- The exact incidence and prevalence of MGRS is unknown, though there is an increasing rate in adults >50 years old
- LCPT is a rarer form of MGRS characterized by light chain deposition in the proximal tubule
- LCPT can present with complete or incomplete Fanconi syndrome resulting in renal wasting of glucose, phosphate, bicarbonate, amino acids, proteins, and uric acid
- Initially, our patient's electrolyte derangements were thought to be secondary to her presenting GI losses and/or home carbonic anhydrase inhibitor
- However, the persistence of electrolyte derangements and NAGMA, along with new unexplained proteinuria led to a higher suspicion of MGRS specifically LCPT

## CONCLUSION

- MGRS should be suspected in patients with monoclonal gammopathy of undetermined significance and new onset of acute kidney injury, proteinuria, and NAGMA
- Renal biopsy should be pursued for suspected MGRS for definitive diagnosis and directed treatment

## REFERENCES

1. K Amaador, Peeters H, Minnema MC, et al. Monoclonal gammopathy of renal significance (MGRS) histopathologic classification, diagnostic workup, and therapeutic options. *PublMed*. 2019;7(7):243-254. Accessed April 10, 2024. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6621137/>
2. Leung N, Bridoux F, Batuman V, et al. The evaluation of monoclonal gammopathy of renal significance: a consensus report of the International Kidney and Monoclonal Gammopathy Research Group. *Nature reviews Nephrology*. 2019;15(1):45-59. doi:<https://doi.org/10.1038/s41581-018-0077-4>
3. Jain A, Haynes R, Kothari J, Khera A, Soares M, Ramasamy K. Pathophysiology and management of monoclonal gammopathy of renal significance. *Blood Advances*. 2019;3(15):2409-2423. doi:<https://doi.org/10.1182/bloodadvances.2019031914>
4. Leung N, Bridoux F, Nasr SH. Monoclonal Gammopathy of Renal Significance. *Ingelfinger JR, ed. New England Journal of Medicine*. 2021;384(20):1931-1941. doi:<https://doi.org/10.1056/nejma1810907>
5. Du J, Hu Z. Systematic review and meta-analysis of the clinical features of MGRS. *BMC nephrology*. 2024;25(1). doi:<https://doi.org/10.1186/s12882-024-03458-5>